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Sexual ambiguity and malformation in Zambia: challenges in surgical management

S NATH, L MUNKONGE

SUMMARY

This article addresses the complexity in diagnosis, gender assignment and management in patients with sexual ambiguity and malformed sexual organs. Between 1984 and 1993, nine children and 10 adult patients with this ailment were treated in the University Teaching Hospital, Lusaka, Zambia.

All children had clitorovaginoplasty and adults had different surgical procedures such as feminisation and masculinisation operations. Methods, means and the manner in which we manage these patients in the midst of a scarcity of expert manpower and sophisticated equipment are discussed. Need for a specialised clinic for better management, teaching and research of this unfortunate and highly sensitive congenital defect has been emphasised.

INTRODUCTION

Children born with ambiguous genitalia need proper investigation, gender assignment and of course expert management. In the developing world, health planners are involved in primary health care delivery and are less concerned about this rare but highly sensitive and important problem. This article addresses the issue of complexity in the management of hermaphrodites, and also the means and the manner in which we treat our patients in the midst of a scarcity of manpower and equipment.

Patients with ambiguous genitalia and malformation of sexual organs attend different clinics at different

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ages. They have not been given the deserved attention, care and proper management. This paper also discusses some of the guidelines in order to streamline their management in our environment.

MATERIALS AND METHODS

The University Teaching Hospital (UTH) is based in the Zambian capital, Lusaka, and is the sole referral hospital. Between 1984 and 1993, over a period of 10 years, 29 patients with ambiguous genitalia were seen. Nineteen children in paediatric plastic and 10 adults in the plastic surgical clinic.

Children.

Infants born with inadequate penile tissue without a scrotum pose a difficult problem in assigning gender. To simplify and overcome this, we used estimation of chromatin Barr Bodies of buccal smear for gender assignment. Seven had negative or abnormal Barr Bodies and were sent to other clinics for further management and three absconded during investigation. Nine patients with positive Barr Bodies were assigned as female children and were admitted for surgical treatment (Table I). Maternal history of siblings, medication and hormonal therapy were noted. Clinical examination of the patient included abdomen, inguinal

Figure I: Before surgery.



canal, scrotum/vulva; and the size of the micropenis/clitoris and gonads were recorded. They had routine and non invasive investigation such as ultrasound to know about pelvic adnexa and were subjected to modified clitorovaginoplasty (Figures I and II).

Operation:- The patient was put in the abducted lithotomy position, cleaned and draped. Five ml of 1:250 000 adrenalinic is injected under the skin of the

Table I: Infant intersex in the University Teaching Hospital, Lusaka, Zambia. (Operated on between 1984 and 1993).

No.	Name	Age	Referral	Clinical Findings	Operations
1.	NN	32M	Paed.	Micropenis/clitoris L. small groin TST	Orchidectomy & CL. vaginoplasty
2.	BM	18M	Direct	Micropenis/clitoris L. big R. small vulva	CL. vaginoplasty
3.	ML	27M	Paed.	Very large/clitoris L. R. large vulva	CL. vaginoplasty
4.	SN	25M	Direct	Micropenis/clitoris L & R small vulva	CL. vaginoplasty
5.	SS	36M	Paed.	Micropenis/clitoris L & R large vulva	CL. vaginoplasty
6.	MZ	21M	Paed.	Micropenis/clitoris L. small scrotal TST	Orchidectomy & CL. vaginoplasty
7.	EZ	25M	Paed.	Micropenis/clitoris L. & R. small vulva	CL. vaginoplasty
8.	VM	27M	Direct	Micropenis/clitoris L. & R. large vulva	CL. vaginoplasty
9.	MM	28M	Paed.	Micropenis/clitoris L. small R. big vulva	CL. vaginoplasty

Age 24M = 24 months.

Referral Paed. = from the paediatric wing of the University Teaching Hospital, Lusaka, Zambia.

Referral Direct = from other provinces of Zambia. CL. vaginoplasty = removal or recession of clitoris and vaginoplasty.

TST = Testis; R = Right; L = Left

Figure II: After surgery.



clitoris/penis, and anal area. Foley's catheter size 8-10 is introduced into bladder as a guide to the urethra. Clitoerectomy or clitoral recession is performed retaining its skin flap. With guidance of catheter in the urethra and finger in rectum, midline space is created with blunt and sharp dissection to the level of pelvic peritoneum. The urethral end is taken out through a stab incision in the skin flap of the clitoris. The distal part of this flap is used for anterior lining, and for posterior lining, an advancement flap raised is from perineum for the neovagina. A finger part of a sterile glove filled with cotton wool is used as a mini condom stent. None of the patients developed fistula or had any urinating problem.

Post operatively, mothers were taught to do regular and graduated dilatation by fingers and plastic test tubes of different calibration. Two patients were lost to follow up but surprisingly seven are attending regularly. Their vaginal depth and width are noted at six monthly intervals. Seven (77,77 pc) have satisfactory depth and width. The depth is satisfactory when ranging between 2,5 and 3,5 cm and the width when the vagina easily accommodates any of the examiner's fingers. Two patients had stenosis where mothers were not careful but by graduated dilatation under general anaesthesia satisfactory depth and width were achieved.

Adults.
During the same period, 10 adult patients with sexual malformation and ambiguity were admitted for management from the adult plastic surgical clinic (Table II). They were either direct referrals or referred

by the gynaecologist. In addition to history and clinical examinations as of the children, wherever indicated, investigations included hormonal estimation of adrenal cortex, FHS, LH, PRL, oestrogen and progesterone. X-ray of pelvis and genito-urinary tract, laparoscopy and laparotomy were also performed in some cases. We have not done hystrosalpingography.

The first two cases SK and DL were true hermaphrodites and were reared as boys. They had laparoscopy and hormonal estimation. In both cases, during laparotomy ovotestis were removed. Later, they had mammectomy to acquire the appearance of a male chest. Follow up was for six years and DL conceded having casual sex.

Cases number three and four were complete female with partial atresia of their vagina. EM had a 1 cm thick septum at the junction of 2 cm deep introitus and upper fifth of the vagina. The result of ultrasound of the abdomen and perineum was confusing, therefore, laparotomy was performed to know the status of intrapelvic organs. Haematocolpos was drained by incising the septum per introitus. After one week, the septum was exercised and local vagina flaps were used to achieve uniform vaginal lumen. CH had atresia of the upper four fifths of the vagina. Partial vaginoplasty with a skin graft was performed. Post operatively she wore vaginal stent for seven months with very good results.

Case number five MB had a short, supple and wide vagina of 7 cm depth. Laparoscopy showed agenesis of uterus and normal ovaries. Due to lack of proper vaginal dilators, she told to make and use condom covered wooden dilators of graduated sizes. Cautious coitus was also encouraged. In six months, she was enjoying painfree coitus with her husband. The last five cases had varied malformations. MM and EB (Figures III and IV) were genotype male, and CM was female pseudohermaphrodite.

DM and BM were cases of Mayer-Rokitansky-Küster-Hauser syndrome. Absence of the uterus and fallopian tubes indicates total failure of mullerian duct development but in this syndrome, a uterus is often present, although rudimentary and is due to the failure of vaginal plate formation which eventuate in vaginal atresia. Fifteen to 40 pc of patients of this syndrome may also have minor to major urinary abnormalities

Table II: Adult sexual ambiguity and malformation in the University of Teaching Hospital, Lusaka, Zambia (Operated on between 1984 and 1993.)

No.	Name	Age	Referral	Clinical Findings	Operations
1.	SK	16Y	Male	Microphallus, L & R small SCR, no TST.	Lap. Gonadectomy, Mammectomy.
2.	DL	18Y	Male	Microphallus, L & R small SCR, no TST.	Lap. Gonadectomy, Mammectomy.
3.	EM	14Y	Female	Thick septum into the lumen of the vagina.	Lap; excision of septum & local flap repair.
4.	CH	14Y	Female	Partial atresia of upper 4/5th vagina.	Lap, and partial vaginoplasty.
5.	MB	21Y	Female	Short vagina. Normal vulva, CLT Rudimentary uterus but normal ovaries.	Laparoscopy and non-surgical vaginoplasty.
6.	EB	22Y	Female	Microphallus; L. SCR normal with TST; R. SCR small and TST in groin.	Excision of penis & both TST; and vaginoplasty.
7.	MM	23Y	Female	Microphallus; L & R.SCR less developed; Groin hypotropted TST.	Excision of penis & both TST; and vaginoplasty.
8.	CM	23Y	Female	Rudimentary uterus & OVR; but CLT was hypertrophied.	Laparoscopy, and recession of CLT & vaginoplasty.
9.	DM	20Y	Female	Rudimentary uterus normal OVR, & CLT; agenesis of vagina.	Laparoscopy, and vaginoplasty.
10.	BM	24Y	Female	Rudimentary uterus normal OVR, & CLT; agenesis of vagina.	Laparoscopy, and vaginoplasty.

GDR.RER = Gender reared and brought up since childhood. Age 16y = 16 years; L = Left; R = Right; SCR = Scrotum. Lap = Laparotomy; TST = Testis; OVR = Ovaries; CLT = Clitoris.

Figure III: MM genotype male with microphallus.

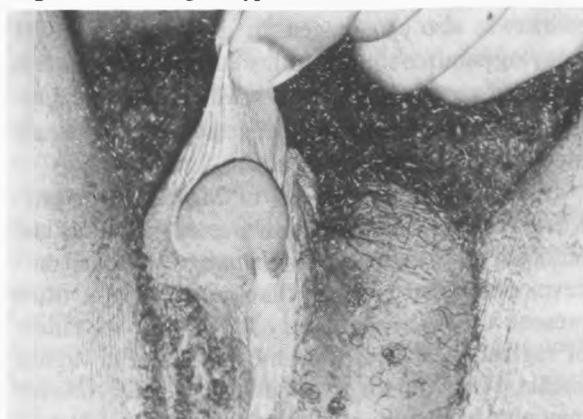


Figure IV:EB genotype male



including congenital absence of a kidney. Twelve patients may have anomalies of their bony skeleton. None of our patients had renal or bony abnormalities. Surgical correction of external genitalia were required in cases number six, seven and eight and all had McIndoe vaginoplasty.¹ Electric dermatome was used for harvesting thick split skin graft from the medial side of the thigh and was sutured inside out over the stent. This was positioned into the space created for the neovagina and sutured around the perineum. The amount of blood collected through the tube into the vacuum drain is carefully monitored in the post operative period. Patients were advised to wear day-time vaginal stent (improvised condom) for at least six months. Out of five complete vaginoplasties two had excellent functional results with distinct serviceable vagina. One had stenosis and was lost to follow up after the first dilatation.

DISCUSSION

The patients with ambiguous genitalia, buccal smear chromosomal Barr Bodies estimation can be positive/negative or abnormal counts. In all positive cases estimation of urinary 17-Ketosteroid will isolate:-

- a. Congenital adrenal hyperplasia (CAH) with elevated level.
- b. Female pseudohermaphrodite – with normal level and with no inguinal or labial gonads but with ovaries in abdomen. All negative cases with abnormal Barr Bodies may need exploration to categorise the type of intersex e.g.:-
- c. True hermaphrodite with ovotestis, occasionally this could be found in patients with positive Barr Bodies as well.
- d. Male pseudohermaphrodite. (Testicular Feminisation or Androgen Insensitivity Syndrome). Normal female breast, genitalia and body habitus. Axillary and pubic hair are sparse. Vagina ends as a blind pouch without cervix. Testis may be intra-abdominal or in groin.
- e. Mixed gonadal dysgenesis (MGD).
- f. Genotype Female with urogenital sinus (UGS).
- g. Genotype Male with bilateral undescended testis and penoscrotal hypospadias.

Ambiguous genitalia has always been a source of fascination to society but only in the last two decades has clinical interest been generated in the management of transsexual patients. There are two distinct groups of

patients who seek treatment. One in their infancy and the other in their adolescence or as an adult. The incidence of an Intersex baby born in Zambia is 1:3 500 births.² This becomes a social emergency and the anguished parents need counselling, explanation of facts and future management. They also need to be informed of the final outcome of the treatment, importance of follow up and the likelihood of secondary surgery in future. The cosmetic effect of clitorovaginoplasty is very gratifying for the parents and the family. We do not do laparotomy or histrosalpingography at this stage. The availability of advance investigating tools like computerised tomographic (CAT) scanner and magnetic nuclear resonance (MNR) imager would have given excellent anatomical information of the intrapelvic organs of the patients with ambiguous abnormal genitalia.³ This information could have been used in the planning of future management. Clitorovaginoplasty on a child is a relatively minor procedure as compared to surgical vaginoplasty on an adult. The possibility of secondary and revisionary surgery on these children would definitely have been obviated had there been the likelihood of having Bicycle Seat treatment⁴ or Frank dilator treatment⁵ in this institution. The Frank dilator is a set of calibrated dilators akin to any other stomal dilators of the body. These are used on the vaginal dimple in the perineum. On the same principle Ingram designed a special bicycle seat mounted on a exercise bicycle. Sustained and regular use of these over the years has created a serviceable neovagina. Nonsurgical vaginoplasty for some selected cases is now an accepted method of treatment.

In the past, we have only treated children with positive Barr Bodies. Others were sent to various clinics for management. None of those children have been referred back to us for exploration. It seems that in the absence of a paediatric endocrinologist, sexual psychologist and overworked urologist and paediatrician these cases must have been lost or neglected. Now, we are admitting all patients of ambiguous genitalia and abnormal sexual organs for treatment and continuous care. The decision of centralising the cases helps in competent treatment, teaching and research. Controversy may arise in doing a feminisation operation in a genotype male child with bilateral undescended testis and penoscrotal hypospadias. The result of masculinising genioplasty is very poor.⁶ In our environment, given the option, parents often request

feminisation operations. One should think very seriously on this issue. Furthermore, psycho-analysis of male hermaphrodites born with ambiguous genitalia has revealed that they would have been happier had they been reared as a woman.⁷

Patients in adolescence and in adulthood present different problems. These patients are brought up either as a boy or a girl. Reassignment of the gender will have a traumatic effect on their social life. In association with the gynaecologist patients are dealt with accordingly. (Table II).

The incidence of the congenital absence of a vagina varies from 1 out of 1 500 to 80 000 births and some of them need creation of a neovagina in future.⁸ Vaginoplasty is one of the most challenging operations in the field of surgery. Dozens of operations have been described. Uses of bowels, rectum, local flaps, gracilis flap and tissues expander have all been reported, but the time-tested McIndoe procedure is still a favoured method of vaginoplasty.⁹

Choosing a stent is a personal choice and bears a very important factor in the successful "take" of the skin graft. Plexiglass, pyrex glass, lucite, balsa wood, dental compound and condoms have been used. Inflatable commercially made stent (Heyer Schulte Co.) are also available. Stents made of wood or glass are very hard and condoms filled with foam with or without vacuum controlled tube are very soft.¹⁰ A firm stent which we make, as follows, is ideal. Dental compound is used to make a penile shaped hollow tube, over which a 1 cm thick foam sheet is sewn. Two large size condoms are unrolled over the foam as if they were being used over the penis. A single tube of a vacuum drain is traversed through this improvised penis and passes out of the tip of the condom.

Creating a 12 to 15 cm deep vagina measured by a calibrated 3.5 cm vaginal mold as a satisfactory result has been reported by a Western author.⁴ A vagina with a 8 cm depth gave painless intercourse.¹¹ A neovagina of this size is not adequate for our community, therefore, we aim to achieve 15 to 20 cm deep and a wider neovagina.

Comment: Management of ambiguous and malformed sexual organs is a multidisciplinary approach. Because of its "Privacy", many hermaphrodites do exist in our society without any treatment. It needs

openness, motivation, eagerness and desire on the part of the patients and empathy, compassion and understanding on the part of the surgeon to achieve the best result.

Patients should be centralised in one special clinic conducted by a plastic surgeon or an interested general surgeon who will keep liaison with other disciplines such as paediatrician, gynaecologist, urologist and endocrinologist. In our series, the services of the pathologist skilled in frozen section were not available when confronted with ovotestis. The availability of CAT scanner or NMR imager would have helped us in firm future planning and avoided many invasive investigations like exploratory laparotomy.

Expert manpower and necessary equipment are needed if we are to help patients with this unfortunate congenital defect.

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